
Memoranda/Mémoires

Strategy for controlling rheumatic fever/ rheumatic heart disease, with emphasis on primary prevention: Memorandum from a Joint WHO/ISFC meeting*

This Memorandum summarizes the report of a meeting held in Geneva on 7–9 September 1994. Experts and representatives from different countries and regions, as well as WHO, the International Society and Federation of Cardiology, UNESCO, and the International Council of Nurses evaluated the experience in controlling rheumatic fever/rheumatic heart disease (RF/RHD) and provided an update on the essential components of RF/RHD prevention, including new areas for research in primary prevention. The meeting's recommendations should be applicable in all countries where RF/RHD is a health problem.

Introduction

Rheumatic fever/rheumatic heart disease (RF/RHD) is the most common cardiovascular disease in children and young adults and remains a major public health problem, especially in developing countries. It results in suffering caused to patients and their relatives, high costs because of repeated hospitalizations (often leading to premature death or disability), and the necessity for increased resources to support the medical and surgical treatment of large numbers of patients (1–6).^{a,b}

* This Memorandum is based on the report of a Joint WHO/ISFC (International Society and Federation of Cardiology) meeting on Rheumatic Fever/Rheumatic Heart Disease Control, with Emphasis on Primary Prevention, which was held in Geneva, Switzerland, on 7–9 September 1994 (unpublished document WHO/CVD/94.1). The participants included 17 temporary advisers, including Professor E.L. Kaplan, USA (Chairman) and Dr R. Talbot, New Zealand (Rapporteur); 3 representatives from other organizations (ISFC, International Council of Nurses, and UNESCO); and 10 members from the WHO Secretariat (Dr P. Nordet (Secretary)). Requests for reprints should be sent to Cardiovascular Diseases Unit, World Health Organization, 1211 Geneva 27, Switzerland. A French translation of this article will appear in a later issue of the *Bulletin*.

^a Report of an Advisory Committee Meeting on streptococcal diseases complex, Geneva, 15–18 November 1983. Unpublished WHO document BVI/STREP/85.1, 1985.

^b A joint WHO/ISFC global strategy to prevent rheumatic fever/rheumatic heart disease. Report of a Consultation, Geneva, February 1990. Unpublished document WHO/CVD/90.3, 1990.

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RF/RHD was a devastating childhood disease in economically developed countries in the 19th century. The decline in RF incidence in these areas was slow but steady after 1900, much more pronounced after the 1940s, and really dramatic in the late 1960s. At present its occurrence is nil or very low, with an incidence below 5.0 per 100 000 population per year, and a prevalence below 0.5 per 1000 schoolchildren (1–3).^b However, the recent resurgence of rheumatic fever and other severe streptococcal infections in some industrialized countries has demonstrated that this is not just a problem in the developing countries, and emphasizes the need for a more intensive approach to public health prevention programmes (1–3).^b

In contrast, RF/RHD was believed to be a rare disease in tropical and subtropical countries during the 19th century. However, since the 1940s it has become a significant health problem, often with very severe effects similar to those observed in Europe a century ago. To date, in developing countries with available data, the RHD mortality rate varies from 0.9 to 8.0 per 100 000 population. Children and young adults still die from acute rheumatic fever; the prevalence in schoolchildren ranges from 1.0 to 10 per 1000 and the incidence from 10 to 100 per 100 000, with a high rate of recurrence and severity. RHD occurs in from 12% to 65% of all cardiac patients and from 1.5% to 4% of all patients admitted to hospital (1–9). In most developing countries, more than 50% of RF/RHD patients are unaware of their disease, and more than 70% do not receive

monthly doses of benzathine penicillin for secondary prophylaxis (1–5, 9–11).^c From these figures, we may conservatively assume that an estimated 12 million people are affected by RF/RHD, with 400 000 deaths annually, and that hundreds of thousands of mainly children and young adults are disabled.

The pathogenetic mechanism of rheumatic fever is still unknown; however, there are two concurrent required factors necessary to initiate the process: a group A beta-haemolytic streptococcal infection of the upper respiratory tract and a specific susceptibility of the individual human host (1–3, 10, 11).^{c,d} Hence, the preventive efforts must be directed towards both these factors.

Pharyngitis or sore throat is common in the community, and occurs throughout the world, especially during childhood. It has been estimated that every child has at least one episode per year and that during endemic conditions Group A beta-haemolytic streptococci (GABHS) can usually be isolated from 20–35% of clinically acute sore-throat cases, in both developed and developing countries. Relatively few individuals (0.3–3%) contract rheumatic fever after acute streptococcal pharyngitis (1–3, 10).^{c,d}

Appropriate case management of symptomatic streptococcal sore throat is important in order to:

- reduce the incidence of suppurative and non-suppurative complications;
- reduce the inappropriate use of antibiotics for upper respiratory infections (when medical information and health education on prevention of rheumatic fever are effective);
- reduce the incidence of symptomatic streptococcal sore throat and the average level of streptococcal antibody in the community;
- reduce the infection contagion rate;
- alter the chain of transmission of GABHS and thus diminish the chance of increasing its virulence.

Overview

WHO has been concerned with RF/RHD prevention and control since 1954. The present meeting was organized to commemorate the 40th anniversary of the very first WHO meeting on this subject (8). Since then, several WHO Expert Committees have directed their efforts towards public health practices

with regard to the study, prevention and control of group A streptococcal infections and RF/RHD (3).^{d,e}

Several programmes centred on secondary prevention—preventing recurrences of rheumatic fever—have been implemented in different countries, including community control of RF/RHD, all of them based on (i) establishment of a rheumatic fever register, (ii) surveillance, (iii) secondary prophylaxis, and (iv) promotion of RF/RHD prevention for physicians, health workers and the general public. These experiences have confirmed the effectiveness and feasibility of a secondary prevention programme. Recently the current WHO Global Programme for the Prevention and Control of RF/RHD, also based on secondary prevention, was successful in most participating countries, thus improving the quality of life in patients and preventing premature disability and mortality. However, it is clear that secondary prevention alone can have only an indirect impact on the number of new cases and on the total number of RF/RHD cases requiring secondary prophylaxis (1–3, 10–14).

Some comprehensive programmes for the prevention of RF/RHD, which integrated primary prevention—early diagnosis and treatment of streptococcal pharyngitis—and secondary prevention into the country's health care system and facilities, have led to significant decreases in mortality, prevalence, incidence, hospital admissions and severity of RF/RHD (1–3, 10, 11, 15–17).

Primary prevention

Populations at risk

School-age children should be the main target of the primary prevention programme. Other high-risk groups (teachers, child care assistants, health workers, etc.) should be included whenever feasible.

GABHS infections

Laboratory aspects. These are:

(a) Optimal management of Group A streptococcal pharyngitis requires laboratory confirmation to ensure that bona fide cases are treated and also to avoid inappropriate use of antibiotics for patients who do not harbour group A streptococci.

(b) Currently in many areas of the world adequate microbiology laboratory facilities are not available. In some such areas, the diagnosis of streptococcal pharyngitis has to be made using defined

^c See footnote a, p. 583.

^d Meeting on the assessment and further development of the WHO programmes on streptococcal diseases and meningococcal infection, Geneva, October 1988. Unpublished document WHO/MIM/STREP/CSM/89.2, 1989.

^e See footnotes a and b, page 583.

clinical criteria and appropriate antibiotics are prescribed without microbiological confirmation.

(c) Laboratory testing is mandatory for specific epidemiological and/or clinical objectives, notably to assess the regional streptococcal epidemiology.

(d) The rapid streptococcal antigen detection tests represent a promising development, allowing the identification of group A streptococci without culture. Since they are expensive and have relatively high specificity and low sensitivity rates, they are unlikely to be used routinely in many countries.

(e) The WHO Collaborating Centres for Reference and Research on Streptococci can assist national public health authorities in the establishment and evaluation of national surveillance and prevention projects for GABHS infections and their sequelae.^f

Diagnosis and management. All classical signs and symptoms of Group A streptococcal pharyngitis or tonsillitis are frequently not present. Clinical diagnosis, therefore, is often uncertain. However, health workers should be aware of the more reliable clinical findings such as exudative pharyngitis or tonsillitis and tender anterior cervical lymph nodes, and whenever possible use them to their advantage. Optimal management ideally requires laboratory confirmation with culture; as an adjunct, direct antigen detection can be used when appropriate.

Penicillin (preferably a single injection of benzathine penicillin G, or alternatively a full 10-day course of oral penicillin V) remains the preferred drug for treating Group A streptococcal respiratory tract infections. In the penicillin-allergic individual a 10-day course of oral erythromycin is the recommended alternative. Tetracyclines and sulfamides should not be used for treatment of Group A streptococcal pharyngitis.

Feasibility under different socioeconomic conditions

(a) Key elements to determine if a primary prevention programme can be initiated include:

- when clinical features are sufficient to make a reasonable diagnosis of streptococcal upper respiratory tract infection which can be documented by adequately trained staff;
- when an adequate public health infrastructure exists; and

— when local human resources are available and can easily be trained for carrying out the programme. Health education is especially crucial.

(b) It is recommended that any primary prevention programme should be part of, or compatible with, national health policy and can be integrated into the national health care system.

Cost-effectiveness

(a) Rheumatic fever/rheumatic heart disease often results in significant mortality and disability, as well as chronic morbidity for children and young adults, manifesting as:

- (i) frequent outpatient visits as well as hospitalization, with the resulting drain on already limited family resources; and
- (ii) loss of educational opportunities.

(b) Medical treatment for rheumatic fever/rheumatic heart disease is not curative and usually has to be continued indefinitely. The costs progressively increase. Surgical treatment is even more expensive and often unaffordable or unattainable.

(c) Prompt treatment of streptococcal pharyngitis prevents rheumatic fever/rheumatic heart disease, as well as other suppurative and non-suppurative complications, and thus obviates the cost of these to families and to society.

Role of physicians, nurses and health workers

Effective organization of human resources for primary rheumatic fever prevention programmes will remain a constant challenge in many developing countries where there is the greatest need. Significant problems include the chronic shortage of medical personnel. Physicians usually assume a directing role and have the important responsibility of educating members of the health team. Appropriate management skills are required for optimal implementation.

Nurses are the most versatile of all health care professionals and are the cornerstone of many disease prevention and control programmes. Their role in the primary prevention of rheumatic fever should be redefined and expanded to include broad responsibility in medical diagnosis and treatment, patients' education, and supervision whenever possible. This committee believes that the involvement and support of the International Council of Nurses (ICN) is crucial to the successful design and implementation of an expanded role for nurses.

Community and village health workers, and rural medical aides, have less training and often function under the supervision of the nurse. How-

^f Further information is available from the Programme on Bacterial, Viral Diseases and Immunology, World Health Organization, 1211 Geneva 27, Switzerland.

ever, they must assume an important role in home visits, education, recording of data, and the referral of sick individuals for advanced medical care when possible.

Health education and information

Health education activities, conducted by doctors, nurses or teachers, can be designed for patients, their relatives, schoolchildren and the general public through lectures, health education sessions, the media (radio, TV, newspapers), posters in schools, health centres and public places, as well as the distribution of booklets and leaflets adapted to the language and sociocultural requirements of the area.

The programme manager and other members of the staff should elaborate and/or disseminate medical information such as: protocol, guidelines, medical information, health education material, etc. to local units and/or personnel participating in the programme. Highest priority must be given to health personnel information and community health education programmes concerning Group A streptococcal pharyngitis and prevention of rheumatic fever and other suppurative and nonsuppurative complications.

These programmes should be aimed at increasing awareness at community level of the importance and feasibility of early diagnosis and treatment of streptococcal pharyngitis, and RF/RHD prevention and control.

New areas for research

Immunology of M protein and development of a safe and effective streptococcal vaccine. Although an anti-streptococcal vaccine remains the hope for the future, there is not yet sufficient evidence to suggest that a safe and effective Group A streptococcal vaccine will be available for mass immunization soon.

Genetic markers to identify people at high risk of developing RF/RHD. There appears to be a genetic influence on the development of rheumatic fever, but exactly which individuals are most susceptible or which Group A streptococci are more likely to initiate the pathogenic process of the disease has not precisely been determined. No genetic marker capable of identifying people at high risk of developing RF/RHD is available yet.

Secondary prevention

Diagnostic criteria

There was consensus that the diagnostic criteria for acute rheumatic fever presented in 1988 by a WHO

Study Group (3) which were emphasized in an American Heart Association medical/scientific statement (7), are still appropriate and do not require modification. Echocardiography is an important diagnostic tool for the evaluation of heart anatomy and performance; however, at present there is insufficient information to document valvulitis in acute rheumatic fever using echocardiography without auscultatory findings.

Secondary prophylaxis

The secondary prophylaxis scheme outlined in 1988 (3) was endorsed. Because of variations in the blood level and the duration of the bactericidal level of penicillin from different brands of benzthine benzylpenicillin, it is recommended that quality control of this antibiotic should be a part of any national programme.

Recommendations

The working group strongly endorsed the following recommendations.

1. A more direct approach to the problem of rheumatic fever and its crippling cardiovascular sequelae should be supported by WHO, by emphasizing primary control measures in addition to the secondary prevention programme.
2. An information document for policy and decision-makers is urgently needed, which includes the goals and objectives of the WHO/ISFC global programme, current information on rheumatic fever/rheumatic heart disease and streptococcal infections, and data regarding the effectiveness and comparative costs of primary and secondary prevention.
3. These recommendations on the prevention of rheumatic fever/rheumatic heart disease (with emphasis on primary prevention) should be implemented by establishing cost-effective and efficient community-based primary prevention rheumatic fever programmes, wherever feasible.
4. To establish and fully implement these public health programmes, it is essential to gain local, national and regional cooperation and support not only from the official government agencies and health planners (and, when appropriate, private industry) but also from the established and potential providers of medical care at the community level (physicians, public health nurses and community health workers). Both health education and the actual delivery of health care should be underlined.
5. Planning for future rheumatic fever control programmes should be carried out in accord with the

specific guidelines elaborated at the present meeting,⁹ with the assistance of WHO.

6. The scientific community should intensify its efforts towards further research for developing a safe and effective anti-streptococcal vaccine, as well as to identify a genetic marker for people at high risk of developing rheumatic fever.

7. The involvement and support of the ISFC and the International Council of Nurses, as well as of national societies, will enhance the implementation of these programmes.

⁹ Joint WHO/ISFC Meeting on Rheumatic Fever/Rheumatic Heart Disease, with Emphasis on Primary Prevention, Geneva, 7-9 September 1994. Unpublished document WHO/CVD/94.1, 1994.

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